PEDIATRIC SURGERY *Update* ©
Vol. 50 No. 03 MARCH 2018

Peliosis

Peliosis hepatis (PH) is a very rare benign disease characterized by multiple small blood-filled cysts of various sizes and shape within the liver parenchyma. Peliosis comes from the Greek word 'pelios' that means reddish or bluish (extravasated blood). What triggers PH is unknown, but dilation of sinusoids might be due to an altered outflow damaging the sinusoid wall and creating dilation of the central vein of the hepatic lobules. Peliosis can be focal or widespread with most cases involving the right hepatic lobe. Peliosis can also occur on other organs such as the spleen, bone marrow, lymph nodes, etc. Etiologic factors associated with PH include drugs, autoimmune mechanisms and infectious causes. Drugs associated with PH include steroids, oral contraceptives, tamoxifen, methotrexate, thiopurine, azathioprine and iron chelators. Alcohol consumption can trigger PH. Imaging studies such as CT-Scan and MRI angiography suggest the diagnosis, but cannot be precised enough since PH cannot be differentiated from adenomas, hemangiomas, focal nodular hyperplasia, Caroli disease or multiple abscess. Lesions are from few millimeters in diameter to 4 cm. Ultrasound may show a pseudocystic lesion of the hepatic parenchyma with intra- or perilesional vascularity. Angiography demonstrates multiple hypervascularized nodules during the late arterial phase with enhancement more pronounced during the parenchymal phase which persists during the venous phase. MRI when combined with hepatospecific contrast material represents the gold standard for radiological diagnosis of PH. Due to the high risk of bleeding an open biopsy using intraoperative US is needed to establish the diagnosis of PH. The disease can cause stenosis of the vena cava when developed in young age. Though mainly asymptomatic, PH can rupture and produce spontaneous hemoperitoneum. Management is usually in the acute setting due to bleeding and consists of either hepatic lobectomy, transplantation or percutaneous embolization.

References:

1- Crocetti D, Palmieri A, Pedulla G, Pasta V, D'Orazi V, Grazi GL: Peliosis hepatis: Personal experience and literature review. World J Gastroenterol. 21(46):13188-94, 2015
2- Hong GS, Kim KW, An J, Shim JH, Kim J, Yu ES: Focal type of peliosis hepatis. Clin Mol Hepatol. 21(4):398-401, 2015
3- Iwata T, Adachi K, Takahashi M: Peliosis Hepatis Mimicking Malignant Hypervascular Tumors. J Gastrointest Surg. 21(6):1095-1098, 2017
4- Dai YN, Ren ZZ, Song WY, Huang HJ, Yang DH, Wang MS, Huang YC, Chen MJ, Zhang JJ, Tong YX, Pan HY: Peliosis hepatis: 2 case reports of a rare liver disorder and its differential diagnosis. Medicine (Baltimore). 96(13):e6471, 2017
5- Tan CHN, Soon GST, Kow WCA: Liver lesions detected in a hepatitis B core total antibody-positive patient masquerading as hepatocellular carcinoma: a rare case of peliosis hepatis and a review of the literature. Ann Hepatobiliary Pancreat Surg. 21(3):157-162, 2017
6- Biswas S, Gogna S, Patel P: A Fatal Case of Intra-Abdominal Hemorrhage Following Diagnostic Blind

Percutaneous Liver Biopsy in a Patient With Peliosis Hepatis. Gastroenterology Res. 10(5):318-321, 2017

Renal Clear Cell Sarcoma

Childhood renal tumors account for 7% of all pediatric cancers. Most cases (90%) are Wilms tumor. Renal Clear Cell Sarcoma (RCCS) is a rare and very aggressive pediatric tumor characterized for its tendency to metastasize to bone often called the bone metastasis renal tumor of children. The term clear cell sarcoma relates to the presence of numerous intracytoplasmic vesicles present in the tumor histology. Other features that differentiate RCCS from Wilms tumor are that they are unicentric in the medullary region of the kidney with foci of necrosis and cyst formation. RCCS is the second most common malignant kidney tumor in children after Wilms tumor comprising approximately 4-6% of all pediatric renal tumors. RCCS is not associated with genetic predisposition syndrome and familial cases have not been reported. RCCS is rare below the age of one year, have a peak incidence between three and five years of age and are more common in males patients. Metastasis from RCCS can also occur to lymph nodes, lungs, liver and brain. RCCS occurs in the same age range as Wilms tumor with no specific radiological features to help distinguish one from the other. Grossly RCCS include large tumor size (more than 10 cm in diameter), mucoid texture, foci of necrosis and prominent cyst formation. Nine histologic different patterns of RCCS have been described. The four important prognostic factors associated with RCCS include treatment with doxorubin, beyond stage I, age at diagnosis greater than two years and tumor necrosis. Management of RCCS at all stages requires aggressive surgical approach followed by chemotherapy and radiotherapy as per NWTS-5 protocols. Overall survival after treatment is 69%. Relapse rates are high and often occur late. Adverse prognostic factors identified are young age, advanced stage IV disease and those with relapse disease.

References:

1- Walke VA, Shende NY, Kumbhalkar DT: Renal Clear Cell Sarcoma - Anaplastic Variant: A Rare Entity. J Clin Diagn Res. 11(1):ED10-ED11, 2017

2- Sinha S, Khurana N, Sarin YK: Clear cell sarcoma of the kidney: report of two cases. APSP J Case Rep. 5(3):32, 2014

3- Brok J, Treger TD, Gooskens SL, van den Heuvel-Eibrink MM, Pritchard-Jones K: Biology and treatment of renal tumours in childhood. Eur J Cancer. 68:179-195, 2016

4- Mandal KC, Mukhopadhyay M, Barman S, Halder P, Mukhopadhyay B, Kumar R: Uncommon renal tumors in children: A single center experience. J Indian Assoc Pediatr Surg. 21(2):61-5, 2016

5- Weaver J, Ho T, Lang A, Koenig JF, Coplen DE, Dehner L, Traxel EJ: Bladder Recurrence of Clear Cell Sarcoma of the Kidney Seven Years After Initial Presentation. Urology. 106:193-195, 2017

6- Wong MK, Ng CCY, Kuick CH, et al: Clear cell sarcomas of the kidney are characterised by BCOR gene abnormalities, including exon 15 internal tandem duplications and BCOR-CCNB3 gene fusion. Histopathology. 72(2):320-329, 2018

Prophylactic Thyroidectomy

Prophylactic removal of all the thyroid gland (total thyroidectomy) is curative treatment for children at risk of developing medullary thyroid cancer (MTC) caused by mutation in

the RET proto-oncogene. Medullary thyroid cancer arises from the parafollicular cells which are responsible for secreting calcitonin. Calcitonin is a sensitive and specific tumor marker for MTC. MTC has an early and high penetrance in hereditary syndrome caused by the RET mutations including multiple endocrine neoplasia (MEN) type 2A and type 2B, and familial MTC which progress to regional lymph nodes and distant metastasis if left unmanageable. The vast majority of MTC in children is hereditary. Children with MEN 2A, MEN 2B and familial MTC can be followed after total prophylactic thyroidectomy with calcitonin levels to monitor for recurrence or development of MTC. Thyroglobulin, a protein precursor of thyroxine produced by the thyroid follicular cells can be a useful test following prophylactic thyroidectomy since many times surgeons leave behind thyroid tissue specially in the nearby region of the recurrent laryngeal nerve (Zuckerkandl tubercle or ligament of Berry). If thyroglobulin is high, an US should be performed to quantify how much residual thyroid tissue was left behind since it has parafollicular cells that can become MTC. Children with MEN2B should go RET genetic analysis and genophenotype ranking or risk level at birth and those with MEN2A and familial MTC before age of five years. Age of prophylactic thyroidectomy recommended include before the first year of age for those MEN children with RET gene mutation and highest genophenotype risk (ATA-D), before age five years for RET mutation and lower risk (ATA-C) and before 10 of age with the minimal risk (ATA-B and ATA-A) or familial MTC. During prophylactic thyroidectomy central lymph node removal is not warranted unless the child has elevated calcitonin level (> 40 pg/mL) with clinical MTC. Annual calcitonin level is needed in all cases and if abnormal thyroidectomy should be performed immediately. The high rate of postoperative hypocalcemia in very young children undergoing prophylactic thyroidectomy has hampered others not to recommend it before the age of three years.

References:

1- Lifante JC, Blanchard C, Mirallia E, David A, Peix JL: Role of preoperative basal calcitonin levels in the timing of prophylactic thyroidectomy in patients with germline RET mutations. World J Surg. 38(3):576-81, 2014

2- Seib CD, Harari A, Conte FA, Duh QY, Clark OH, Gosnell JE: Utility of serum thyroglobulin measurements after prophylactic thyroidectomy in patients with hereditary medullary thyroid cancer. Surgery. 156(2):394-8, 2014

3- Starenki D, Park JI: Pediatric Medullary Thyroid Carcinoma. J Pediatr Oncol. 3(2):29-37, 2015

4- Kluijfhout WP, van Beek DJ, Verrijn Stuart AA, Lodewijk L, Valk GD, van der Zee DC, Vriens MR, Borel Rinkes IH: Postoperative Complications After Prophylactic Thyroidectomy for Very Young Patients With Multiple Endocrine Neoplasia Type 2: Retrospective Cohort Analysis. Medicine (Baltimore). 94(29):e1108, 2015

5- Boybeyi-Turer O, Vuralli D, Karnak I, Gonc N, Yalcin E, Orhan D, Kandemir N, Tanyel FC: Surgical and clinical strategies in the management of thyroid medullary carcinoma in children with and without ret proto-oncogene mutations. Turk J Pediatr. 58(4):436-441, 2016

6- Bussieres V, Roy S, Deladoey J, Rousseau E, St-Vil D, Piche N: Prophylactic thyroidectomies in MEN2 syndrome: Management and Outcome. J Pediatr Surg http://dx.doi.org/10.1016/j.jpedsurg.2017.11.015

***Edited by: Humberto Lugo-Vicente, MD, FACS, FAAP**

Professor of Pediatric Surgery, University of Puerto Rico - School of Medicine,

Rio Piedras, Puerto Rico. Director - Pediatric Surgery, San Jorge Childrens Hospital.
Address: P.O. Box 10426, Caparra Heights Station, San Juan, Puerto Rico USA 00922-0426.
Tel (787)-999-9450 E-mail: *titolugo@coqui.net*
Internet: http://home.coqui.net/titolugo

ISSN 1089-7739